Review
JCAM

Signs on Obstetric Ultrasound Images in the Second Trimester

İkinci Trimester Obstetrik Ultrasonografik Görüntülemede İşaretler

Signs on Obstetric Ultrasound

Fatma Uysal
Çanakkale Onsekiz Mart University, Department of Radiology, Çanakkale, Turkey

Özet

Bir radyolojik işaret sıklıkla benzer bir grup patolojiyi düşündüren belirli bir nesneye benzetilir. Bu tür benzerlikler farkındalığı artırmada ve ayırıcı tanı listesinin kısaltılmasında yararlıdır. Radyolojinin diğer alanlarında olduğu gibi obstetrik ultrasonografide de tanıyı kolaylaştırmak için birçok işaret tanımlanmıştır. Prenatal ultrasonografide saptanan bu işaretler spina bifida'da görülen limon ve muz işareti, korpus kallozum agenezisinde görülen gözyaşı damlası işareti, Dandy Walker sendromunda ve posterior üretral valvde görülen anahtar deliği işareti, duodenal atrezide görülen double bubble işareti, yarık dudak olgularında görülen tavşan dudak işareti, ekstremite anomalilerinden kaudal regresyon sendromunda görülen kurbağa bacağı işareti, sandal gap deformitesi görünümleridir. Bu işaretleri içeren olgulara ait 2 ve 3 boyutlu ultrasonografi görüntüleri benzetildikleri nesne şekilleri eşliğinde sunulacaktır.

Anahtar Kelimeler

Obstetrik Ultrasonografi; İşaretler; Fetal Anomali

Abstract

A radiologic sign resembles a specific object, often suggesting a group of similar pathologies. These types of similarities are useful to increase awareness and shorten the list of differential diagnoses. As in other fields of radiology, in obstetric ultrasonography many signs have been described to ease diagnosing. These signs identified on prenatal ultrasonography include the lemon and banana signs observed in spina bifida, teardrop sign in corpus callosum agenesis, keyhole sign in Dandy-Walker syndrome and posterior urethral valve, double bubble sign in duodenal atresia, rabbit lip in cleft lip cases, and in extremity anomalies, frog leg sign in caudal regression syndrome and sandal gap sign. The 2- and 3-dimensional ultrasound images of cases with these signs accompanied by the objects they resemble are presented in this review.

Keywords

Obstetric Ultrasound; Signs; Fetal Anomaly

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Corresponding Author: Fatma Uysal, Çanakkale Onsekiz Mart University, Department of Radiology, Çanakkale, Turkey.
GSM: +905052874231 E-Mail: afatmauysal@gmail.com

Introduction

In radiology practice some images that are similar to described signs can be useful to classify similar diseases. These signs shorten the list of differential diagnoses and may help doctors reach a final decision for the patient. This report presents the 2- and 3-dimensional ultrasonography images of 9 important and frequently observed radiological signs described on obstetric ultrasound together with sketches and pictures.

Lemon sign

Diagnosis of neural tube defects requires experience and careful examination. In spite of careful examination small open neural tube defects may be missed. As a result cranial anomalies are used as a marker for the presence of spina bifida. A symmetrical flattened lemon sign in the ventral part of the cranium (Fig. 1) shows with neural tube defects and is used as an indicator of spinal opening. The lemon sign is very helpful before 24 weeks gestation in diagnosis of spina bifida in high-risk population; however as gestational age advances as a result of development of the fetus the lemon sign becomes unclear and reliability reduces [1,2]. The lemon sign is not specific to spina bifida. It may accompany encephalocele, Dandy-Walker malformation with encephalocele, cystic hygroma, diaphragmatic hernia, corpus callosum agenesis, fetal hydrops, umbilical vein varices and double-vein cord anomalies. When the lemon sign is present cranial findings such as ventriculomegaly, microcephaly, obliteration of the cisterna magna, compression of the cerebellar hemispheres and ventral-directed orientation (banana sign) should be investigated and the vertebral column requires careful evaluation [3,4].

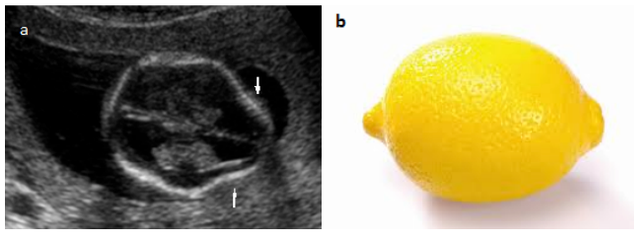

Figure 1. Lemon and banana signs (spina bifida) belonging to a 17-week fetus(A,B) (2A,C) B-mode axial through cranium(1A,2A), 3-D coronal image through vertebral column)(2C), Biconcavity of frontal bones in fetal neurocranium (lemon sign)(1A), lemon(1B)

Banana sign

A sign of Chiari II malformation, as a result of downward migration within the posterior fossa of the fetus the cerebellum wraps tightly around the brain stem and obliterates the cisterna magna (Fig. 2). The cerebellum gains the appearance of a banana. Additionally observed in the majority of spina bifida fetuses, this finding is lost after 24 weeks. This sign may be rarely observed in normal fetuses. Frequently there is accompanying hydrocephaly [5,6].

In risk populations before 24 weeks spinal defects on US are small and thus difficult to see, increasing the importance of visualizing indirect findings of spina bifida. The hemispheres wrap the brain stem and gain a "C" shape (banana sign). On longitudinal sonography just as open spinal and skin defects of spina bifida may be seen, expansion of the spinal canal diameter and increased interpeduncular distance may be identified [3].

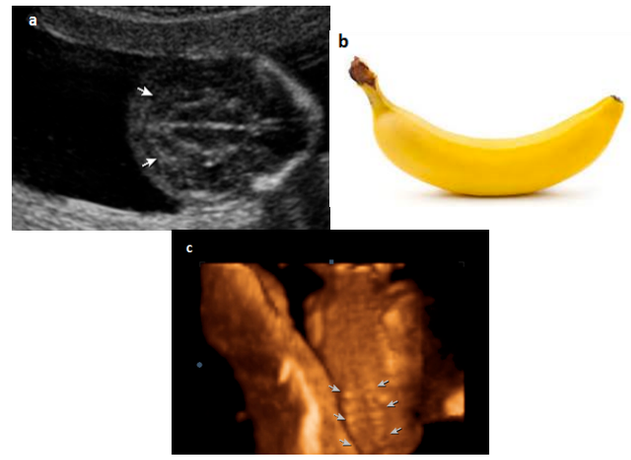

Figure 2. Banana sign in the cerebellum and obliteration of the cisterna magna(A), banana (B), Wide spina bifida appearance on 3-D image in the lumbar region(C).

Teardrop sign

Corpus callosum agenesis (CCA) is lack of development in varying degrees of the caudal part of the corpus callosum (corpus and splenium). The corpus callosum completes development between the 12th and 18th weeks of pregnancy. In CCA the cavum septum pellicidum (CSP) is not found. CCA may be complete or incomplete.

The incidence in the population is between 0.3-0.7%, and is 2-3% in patients with developmental disorders [7,8]. The etiology varies. Generally it develops due to genetic factors. It shows autosomal dominant, autosomal recessive and X-linked inheritance [9]. Additional anomalies are present in 50% of cases. The most frequent are Dandy-Walker malformation and congenital heart anomalies. Abnormal karyotype (trisomy 18 and 8) are observed at a rate of 20% [10].

Prenatal diagnosis should be suspected in situations of atrium widening and CSP not being observed. Widening of the atrium and occipital horns on axial plane and separation of the body of the lateral ventricle forms a "teardrop" appearance which is specific for diagnosis and frequently found (Fig. 3). As a result of more definite widening at the level of the occipital horn of the lateral ventricles the so-called teardrop configuration (colpocephaly) is observed. The most consistent and identifiable sign is the small appearance of the choroid plexus which appears as a teardrop-shaped pendant (Fig. 3). To confirm CCA coronal or sagittal plane images of cases with dilated atrium and teardrop sign should be taken. Findings such as widening

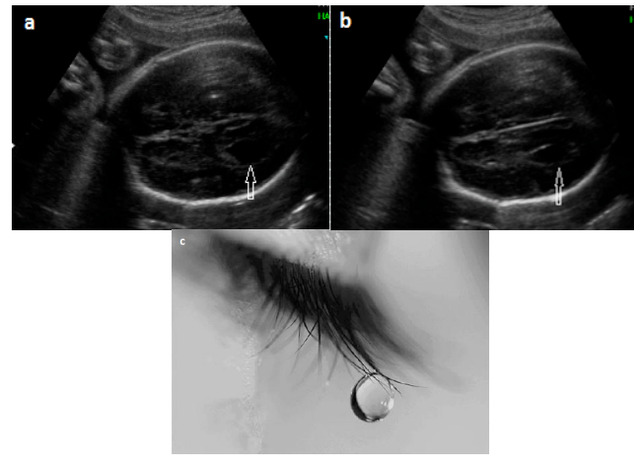

Figure 3. Teardrop sign (corpus callosum agenesis) (A,B), Thirty-week fetus axial section at lateral ventricular level in the cranium showing corpus callosum agenesis with teardrop sign in the lateral ventricle occipital horn, tear drop(C)

of the interhemispheric fissure and third ventricle higher than normal are frequently found. Color Doppler ultrasound showing abnormal branching of the anterior cerebral artery is a supporting finding for diagnosis. In the incomplete type the "teardrop" sign may be present. However other findings may not be clear. As a result, prenatal diagnosis is difficult. In patients with prenatal diagnosis fetal karyotyping should definitely be completed. While the prognosis is not known for definite, isolated CCA with normal or near-normal development of intelligence has been identified. Some cases are only suspected on US and prenatal MRI is recommended for diagnosis. In the majority of cases diagnosis can be made with MRI [11].

Keyhole sign (Dandy-Walker malformation)

Dandy-Walker malformation (DWM) and Dandy-Walker variant (DWV) are non-specific congenital brain malformations as a result of cerebellum and vermis development anterior-superior in the rhombencephalon beginning in the ninth week of the embryonic period and not completing by the 16-17th week of pregnancy. It is reported both isolated and as a component of many dysmorphic pathologies. DWM has an incidence of 1/25,000-35,000 in live births. The incidence of Dandy-Walker variant is thought to be higher and it is reported to form one third of posterior fossa lesions. Ultrasound findings of classic DWM include; larger than normal cisterna magna and/or posterior fossa cyst (cystic dilatation of the fourth ventricle), full or partial agenesis of cerebellar vermis, hydrocephalus or partial expansion of the atriums. In the Dandy-Walker variant ultrasound findings are similar to DWM, though differences include a possibly smaller posterior fossa cyst and varying degrees of agenesis of vermis (full or partial absence of the lower lobe of the vermis). Intracranial ventricular dilatation may or may not accompany this syndrome. The absence of the lower lobe of the vermis creates the "keyhole" observed ultrasonographically which is very useful in DWV diagnosis (Fig. 4) [12,13].

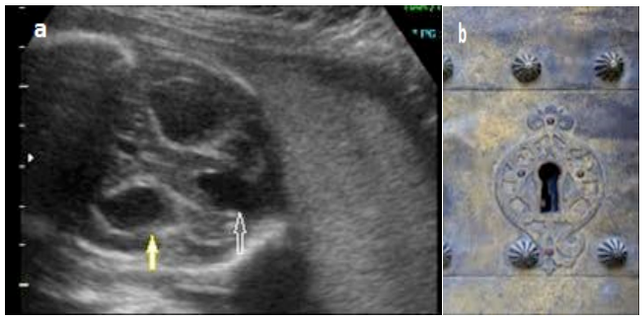

Figure 4. Keyhole sign (Dandy-Walker malformation-variant). Twenty-seven week fetus, axial image at cerebellar level in the cranium with cystic appearance in the posterior fossa related to the 4th ventricle (keyhole sign (empty arrow))(A), dilatation in the lateral ventricle occipital horn (yellow arrow), Keyhole (B).

Keyhole sign (posterior urethral valve)

Typical presentation of PUV during routine prenatal ultrasound is identification of hydronephrosis.

The large rate of PUV cases identified on prenatal sonography with no evidence identified after birth is very surprising [14,15]. On routine second trimester obstetric ultrasound the PUV anomaly may frequently be accompanied by bilateral hydroureteronephrosis thickening of the bladder wall and in males the keyhole sign at the bladder neck (Fig. 5). This situation forms 10% of all prenatally identified hydronephrosis [14-16].

The keyhole sign is considered to be very specific to PUV with

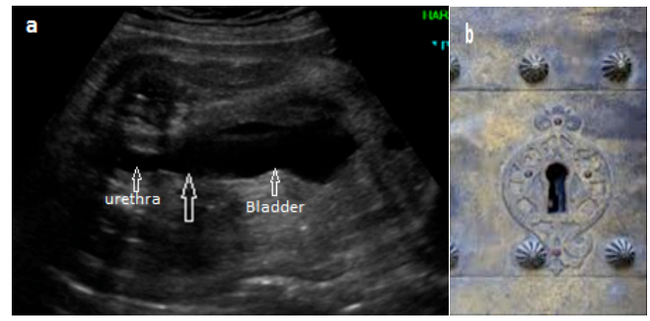

Figure 5. Keyhole sign (posterior urethral valve). Nineteen-week fetus, widening (arrow) of the bladder connection of the urethra on coronal image at the bladder level(A), keyhole(B)

findings of dilatation of the posterior urethra with posterior urethral obstruction [17].

Bernardes et al. [18] in their publication of a series of classic prenatal US found the sensitivity of the sign was high however the specificity was low. The best diagnostic markers were increased thickening of the bladder wall and dilatation of the bladder. The keyhole sign is not regarded as a reliable marker for PUV.

Double bubble sign

In prenatal diagnosis of duodenal atresia, US, karyotype analysis and fetal echocardiography are used. On ultrasonography when the fetal abdomen is shown in the transverse plane the first section of the duodenum, which dilates with the stomach, is full of fluid, showing two separate cystic structures related to each other (double bubble finding) which is used in diagnosis (Fig. 6). This finding is analogous to the gas-filled double-bubble found on radiologic investigation of newborns with duodenal atresia [19]. The double-bubble finding is identified on average at 24 weeks, but it is reported to be identified in earlier weeks [20]. Petrikovsky et al. [21] presented the case of a successful duodenal atresia diagnosis made in the 14th week of pregnancy.

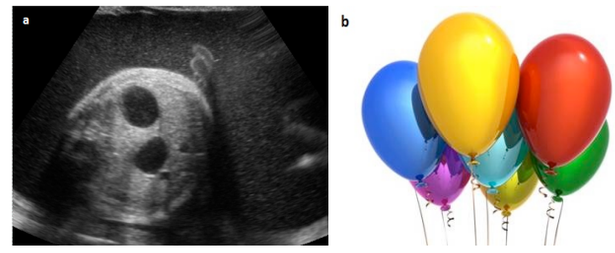

Figure 6. Double bubble sign (duodenal atresia). Fifteen-week fetus, double bubble sign due to stomach and duodenum dilatation on axial section at the stomach level(A), balloons(B)

Rabbit lip sign

In the second trimester, performing a detailed US to identify fetal anomalies in all pregnancies has become a basic application. Parallel to the increase in technology and knowledge more fetal anomalies are able to be diagnosed in the prenatal period. Cleft lip/palate (Fig. 7) is one of the most common congenital anomalies with an incidence of 1 per 1000 live births [22]. In Turkey the incidence of cleft lip/palate is 0.95 per thousand with isolated cleft lip incidence of 0.77 per thousand [23]. Cleft lip/palate may be isolated but may also accompany chromosomal, structural anomalies and nearly 350 syndromes [24].

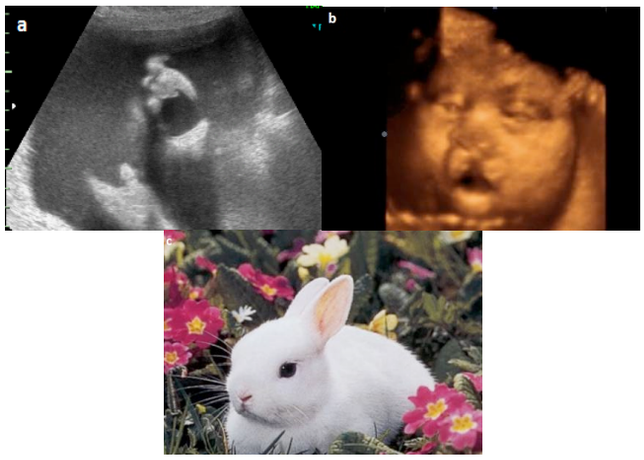

Figure 7. Rabbit lip sign (cleft lip). Thirty-three week fetus, B-mode image of the lips(A), 3-D image of the face with appearance appropriate for upper cleft lip(B), rabbit(C)

The structures on the midline of the fetal face fully meld by the 7th week of pregnancy. However cleft lip/palate diagnosis may not be possible with high accuracy until the 13-14th week of pregnancy [25]. Recently the use of 3- and 4-dimensional ultrasonography has increased the diagnosis rate of facial defects [26]. Johnson et al. [27] showed the use of 3-dimensional ultrasound had increased the diagnosis rate from 48% to 76%.

Frog leg sign

Caudal regression syndrome (CRS) is a rarely-observed congenital anomaly with a variety of degrees of early gestational developmental disorders. This situation is known as sacral agenesis or caudal dysplasia. This malformation is thought to be caused by neuralization defects forming around the 28th day of pregnancy. While maternal uncontrolled diabetes, genetic predisposition and vascular hypoperfusion are possible risk factors the true pathogenesis is unclear. CRS diagnosis is generally given prenatally however again a variable number of newborns with varying degrees of anomalies may be diagnosed. When shorter CRL than expected according to last date of menstrual period and incomplete vertebral ossification on both grayscale and 3-D ultrasonography images are identified, caudal regression syndrome should be considered (Fig. 8). Flexion and immobility of both lower extremities are among the observed findings [28].

Characteristic US findings include sudden interruption to the spine and abnormal positioning of the lower extremities. The femur bones are typically in a fixed V position, appearing like a typical "Buddha pose". Scanning for possible accompanying urinary and intestinal malformations should be completed [29].

Sandal gap sign

The sandal gap deformity is a wider than normal space between the first and second toes and includes medial displacement of the big toe (Fig. 9). The separation of the big toe is reported in 33.3% of Down syndrome cases [30]. However it is not a typical finding for other syndromes.

This deformity is observed in many normal fetuses or neonates as a variant of normal. If prenatal ultrasonography shows sandal gap deformity careful scanning for other risk factors of Down syndrome is necessary.

If prenatal ultrasonography shows no other abnormalities including clinical risk factors the separation of the big toe should be accepted as a normal variant [31].

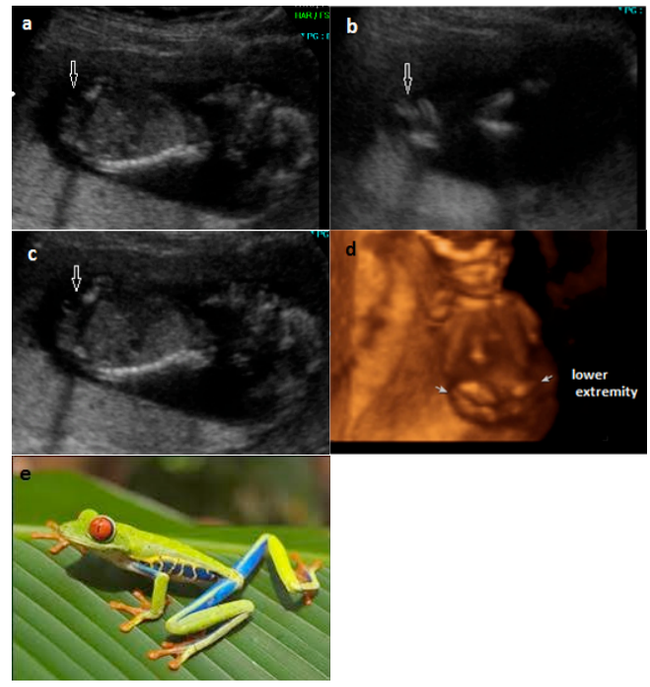

Figure 8. Frog leg sign (caudal regression syndrome). Sixteen-week fetus, consecutive sagittal B-mode images(A,C), 3-D image showing absence of sacrum and both lower extremities in flexion position(D), frog's legs(E)

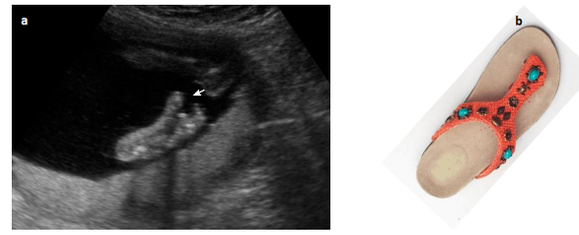

Figure 9. Sandal gap sign (Down syndrome). Eighteen-week fetus, B-mode coronal image including sole of foot with increased distance between 1st and 2nd toe (arrow)(A), sandal(B)

Conclusion

Described signs with similarity to objects or shapes on obstetric ultrasonography, as in other fields of radiology, carry great importance to ease diagnosis. As a result it is necessary to know the signs and the main pathologies that cause them.

Competing Interests

The authors declare that they have no competing interests.

References

1. Thomas M. The lemon sign. Radiology 2003;228(1):206-7.
2. Sebire NJ, Noble PL, Thorpe JG, Snijders RJ, Nicolaides KH. Presence of the lemon sign in fetuses with spina bifida at the 10-14 week scan. Ultrasound Obstet Gynecol 1997;10(6):403-5.
3. D'Addario V, Rossi AC, Pinto V, Pintucci A, Di Cagno L. Comparison of six sonographic signs in the prenatal diagnosis of spina bifida. J Perinat Med 2008;36(4):330-4.
4. Nicolaides KH, Campbell S, Gabbe SG, Guidetti R. Ultrasound screening for spina bifida: cranial and cerebellar signs. Lancet 1986;12:2:72-4.
5. Benacerraf BR, Stryker J, Frigoletto FD., Jr Abnormal US appearance of the cerebellum (banana sign): indirect sign of spina bifida. Radiology 1989;171(1):151–3.
6. Van den Hof MC, Nicolaides KH, Campbell J, Campbell S. Evaluation of the lemon and banana signs in one hundred thirty fetuses with open spina bifida. Am J Obstet Gynecol 1990;162(2):322–7.
7. Grogono JL. Children with agenesis of the corpus callosum. Dev Med Child Neurol 1998;10(5):613-6.
8. Jerret JS, Serur D, Wisniewski K, Fisch C. Frequency of the agenesis corpus callosum in the developmentally disabled population as determined by computerized

tomography. Pediatr Neurosci 1986;12(2):101-3.
9. Young ID, Trounce JQ, Levene MI, Fitzsimmons JS, Moore JR. Agenesis of the corpus callosum and macrocephaly in siblings. Clin Genet 1985;28(3):225-30.
10. Parrish M, Roessman U, Levinsohn M. Agenesis of the corpus callosum: A study of the frequency of associated malformations. Ann Neurol 1979;6(4):349-54.
11. Pilu G, Sandri F, Perolo A, Pittalis MC, Grisolia G, Cocchi G,et al. Sonography of fetal agenesis of the corpus callosum: A survey of 35 cases. Ultrasound Obstet Gynecol 1993,3(5):318-29.
12. Keogan MT, De Atkine AB, Hertzberg BS. Cerebellar vermian defects: antenatal sonographic appereance and clinical significance. J Ultrasound Med 1994; 13(8): 607-11.
13. Chang MC, Russel SA, Callen PW, Filly RA, Goldstein RB. Sonographic detection of inferior vermian agenesis in Dandy-Walker malformations: prognostic implications. Radiology 1994;193(3):765-70.
14. Lopez Pereira, Martinez Urrutia MJ, Jaureguizar E. Initial and long-term management of posterior urethral valves. World J Urol 2004;22(6):418–24.
15. Agarwal S. Urethral valves. BJU Int 1999;84:570–78.
16. Yohannes P, Hanna M. Current trends in the management of posterior urethral valves in the pediatric population. Urology 2002;60(6): 947–53.
17. Mahony BS, Callen PW, Filly RA. Fetal urethral obstruction: US evaluation. Radiology 1985;157(1):221–4.
18. Bernardes LS, Aksnes G, Saada J, Masse V, Elie C, Dumez Y, et al. Keyhole sign: how specific is it for the diagnosis of posterior urethral valves? Ultrasound Obstet Gynecol. 2009;34(4):419-23.
19. Takeshita T, Nomura Y, Nakamura Y. Antenatal diagnosis of duodenal atresia using ultrasonic scanning. Asia Oceania J Obstet Gynaecol 1982;8(3):269-73.
20. Romero R, Ghidini A, Costigan K, Touloukian R, Hobbins JC. Prenatal diagnosis of duodenal atresia: does it make any difference? Obstet Gynecol 1988;71(5):739-41.
21. Petrikovsky BM. First-trimester diagnosis of duodenal atresia. Am J Obstet Gynecol 1994;171(2):569-70.
22. Wyszynski DF, Beaty TH. Review of the role of potential teratogens in the origin of human nonsyndromic oral clefts. Teratology 1996;53:309-17.
23. Tunçbilek E. Türkiye'de konjenital malformasyon sıklığı, dağılımı, risk faktörleri ve yenidoğanların antropometrik değerlendirilmesi. Ankara:Tubitak Matbaası,1996: 94.
24. Gorlin RJ, Cervenka J, Pruzansky S. Facial clefting and its syndromes. Birth Defects Orig Artic Ser 1971;7(7):3-49.
25. Özçimen EE, Üçkuyu A, Üstüner I, Kayhan İ, Yanık FF. Antenatal diagnosis of cleft lip and palate by 2-D and 3-D utrasound, case report and review of the literature. Türkiye Klinikleri (J Gynecol Obst) 2006;16: 98-101.
26. Uysal F, Uysal A, Güngör AC, Coşar E. True cyclopia and proboscis: an interesting prenatal condition with normal nose. J Clin Anal Med 2013; DOI: 10.4328/JCAM.1588.
27. Johnson DD, Pretorius DH, Budorick NE, Jones MC, Lou KV, James GM, et al. Fetal lip and primary palate: Three-dimensional versus two-dimensional us. Radiology 2000;217(1):236-9.
28. Singh SK, Singh RD, Sharma A. Caudal regression-case report and review of the literature. Pediatr Surg Int 2004;21(7): 578-81.
29. Bashiri A, Sheizaf B, Burstein E, Landau D, Hershkovitz R, Mazor M. Three dimensional ultrasound diagnosis of caudal regression syndrome at 14 gestational weeks. Arch Gynecol Obstet 2009;280(3):505-7.
30. Azman BZ, Ankathil R, Siti Mariam I, Suhaida MA, Norhashimah M, Tarmizi AB, et al. Cytogenetic and clinical profile of Down syndrome in Northeast Malaysia. Singapore Med J 2007;48(6):550-4.
31. Wilkins I. Separation of the great toe in fetuses with Down syndrome. J Ultrasound Med 1994;13(3):229–31.

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